Sickle Cell Disease

An Overview of Current Services and Emerging Needs in the Commonwealth

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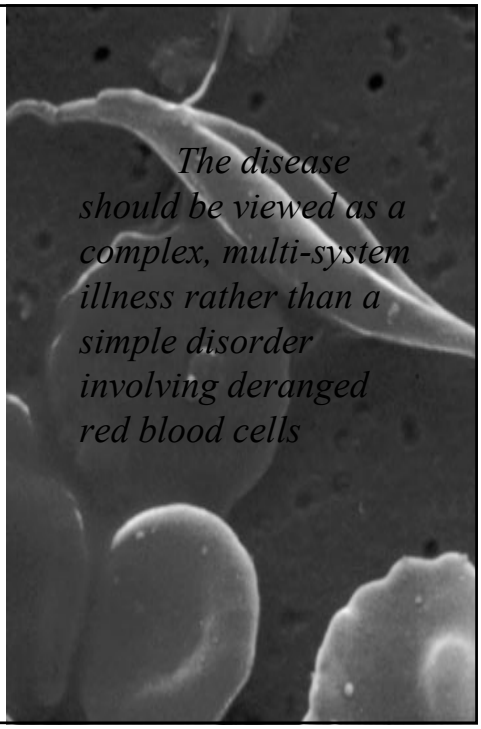
Presentation Objectives

- Describe sickle cell disease and its complications
- Review the impact of sickle cell disease
- Give examples of state and community-based efforts to improve the quality of life for individuals and families effected by sickle cell disease
- Identify resources needed to meet the medical and social needs of effected individuals and families

What is Sickle Cell Disease?

- Sickle Cell Disease (SCD) is a collective term used to describe a spectrum of genetic disorders that affect the shape and function of the red blood cell
- Red Blood Cells (RBCs) form crescent or sickle shape and slow or block blood flow
- Constant breakdown of damaged RBCs releases proteins that increase inflammation, clotting, and cause vasoconstriction
- Results in extreme pain and chronic damage to multiple organs



Complications from Sickle Cell Disease

- Chronic hemolytic anemia
- Acute splenic sequestration-spleen traps RBCs
- Aplastic crisis-bone marrow stops making RBCs
- Painful crises (bones and chest)
- Bacterial infections (children)
- Acute chest syndrome (children)
- Kidney failure
- Gallbladder stones and inflammation
- Avascular necrosis
- Pulmonary hypertension
- Stroke (children)

Medical Management

- Prevent bacterial infections
  - Immunizations
  - Penicillin treatment from 3 mos. to 5 yrs.
- Transfusions
  - To prevent stroke, decrease crises, reduce complications, correct anemia
- Comprehensive care
  - Comprehensive Pediatric and Adult Sickle Cell Centers
  - Specialist consultation
  - Hydroxyurea (anti-sickling agent taken daily)
  - Pain medication, hydration, oral chelation, oxygen
  - Dialysis or kidney transplant
  - Surgery for gallbladder removal
  - Hip/shoulder replacement for avascular necrosis
  - Bone marrow transplant
- Future treatments
  - Genetic engineering

Hydroxyurea

- 50% reduction in crises
- 40% reduction in mortality
- If given routinely:
  - Up to 70 lives could be saved every 10 years
  - Up to $13.1 million in Medicaid payments could be saved every 10 years

Supportive Services

"Teaching the skills necessary for coping with this disease should begin at the time of diagnosis and continue throughout the life of the patient."

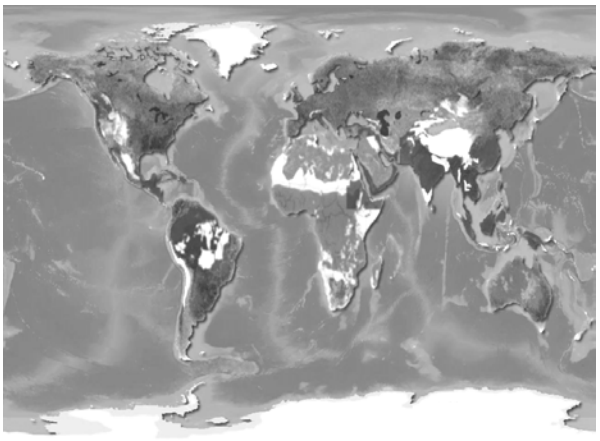
Source: NIH, The Management of Sickle Cell Disease, 1984

- Pain management skills
- Ongoing education
- Community-based supportive services
- School and work interventions
  - IEP
  - Accommodations specific for SCD
  - Career/vocational planning

Impact of Sickle Cell Disease

~Two million people living with SCD worldwide

- Africa
- South and Central America
- Caribbean
- Mediterranean
- India
- Saudi Arabia



Domestic Statistics

No national registry for SCD

- ~80,000 living with SCD in the United States
- 1 in 375 African Americans
- 1 in 1100 to 1400 Hispanics
- 1 in 58,000 Caucasians
- > 2 million Americans have sickle cell trait

Source: National Heart, Lung and Blood Institute (NIH, 2007)
Smith LA et al. Pediatrics 2006

Health Burden of SCD

- Nationally,the number of hospitalizations among adults with SCD in 2004 was **83,149**
- Virginia = 1732
- The total hospital costs in 2004 for hospitalizations principally for SCD were approximately **$488 million**

- Among those hospital stays principally for SCD
  - 66% were paid by Medicaid
  - 13% were paid by Medicare
  - 21% private insurance

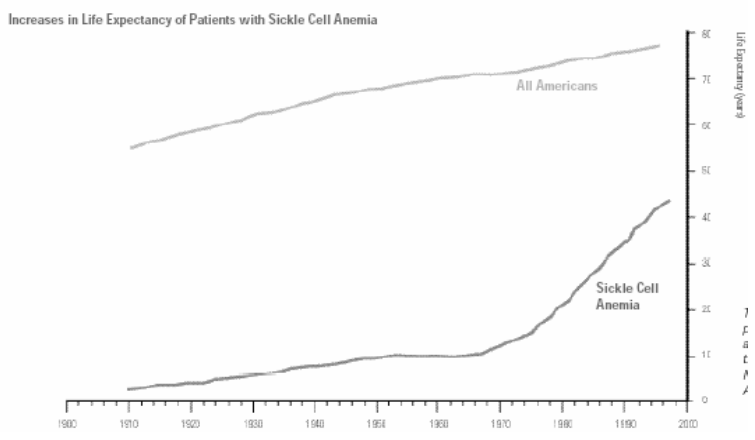
Source: Healthcare Cost and Utilization Project (HUP), ARHQ - 2004

Life Expectancy

- In the US, as recently as 1970, the average person with SCD died in childhood
  - 10% before their 10th birthday
  - 50% before 21st birthday
- In 2000, as a result of early detection and improved treatment, sickle cell patients live into their 40s and 50s

Source: National Institutes of Health

Life Expectancy for Sickle Cell Patients



Sickle Cell Services in the Commonwealth

- *The Virginia Sickle Cell Awareness Program*
- *The Virginia Newborn Screening Program*
- *Pediatric Comprehensive Sickle Cell Clinics*
- *Community-based Sickle Cell Programs*

Sickle Cell Disease in Virginia

- **~3,700** living with SCD
  - **1 in 325** African Americans (8% higher than national average)
  - **< 1%** identified in other ethnic groups
  - **1 in 12** African Americans identified with sickle cell trait
- **~ 75 newborns** identified with disease yearly

Source: Virginia Birth Records, Virginia Sickle Cell Awareness Program Newborn Screening Tracking Database

Virginia Sickle Cell Awareness Program (VASCAP)

Code of Virginia 32.1-68.

- Title V ($125,000) funded statewide program for the education and voluntary screening of individuals for sickle cell disease, trait and other related hemoglobinopathies.
- Located at the Virginia Department of Health, Division of Women's and Infants" Health
- 1FTE

VASCAP Services

Screening Services

- ~9000 adults screened yearly
  - Family Planning
  - Maternity clinics
  - Newborn family studies (parent and siblings)
  - Limited, targeted community-based screening

VASCAP Services

Health education and promotion

- Community groups
- Churches
- Health care providers
- Genetic counselors
- Health department personnel
- Medical, nursing and social work students
- School personnel
- Individuals living with SCD
- Collaboration with regional, national and international partners

Contract management

- Pediatric Comprehensive Sickle Cell Clinics
- Community-based programs

Newborn Screening Services

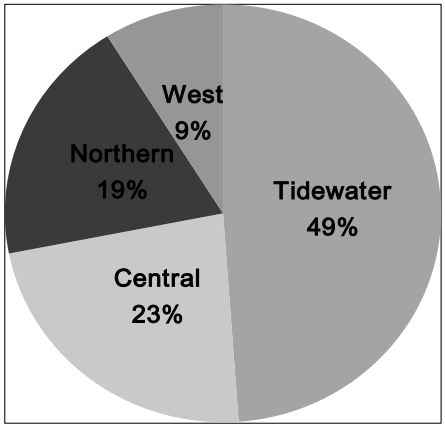
Code of Virginia 32.1-65 through 32.1-69
Sickle cell screening and treatment

- In July of 1989, Virginia began screening all newborns for SCD
- **Today, 1257** newborns have been identified with SCD

VASCAP collaborates with Virginia Newborn Screening Program to provide:

- Tracking of newborns identified with SCD to insure early entry to care
- A database of all newborns identified with SCD through the newborn screening program
- Direct parent notification of newborn carrier status (> 2000 yearly) and telephone counseling and or referral upon request

Where are these families located?



Eastern Virginia
Children's Hospital of the King's Daughters: 534 Clients (49%)
Portsmouth Naval Hospital (~75)

Central
VCU Health System - Richmond
251 Clients (23%)

Northern Virginia

INOVA Fairfax - Falls Church
204 Clients (19%)

West
UVA – Charlottesville
102 Clients (7%)
Roanoke Carilion (2%)

Enhancing Access to Care

Pediatric Comprehensive Sickle Cell Clinic Network

General Funds

1994

- Yearly allocation $250,000 to develop regionally located sickle cell clinics and support community-based sickle cell programs

2007

- As a result of a 136% increase in the number of clients receiving services the GA increased funding to $450,000
- And then decreased to $350,000

- Provides culturally competent comprehensive medical and support services that are:
  - collaborative
  - family-centered
  - and community-based
- Four clinics are currently serving 1091 clients between between the ages of 1 mo. and 22 years
- 3794 patient visits during FY 06

State Funded Pediatric Comprehensive Sickle Cell Center Catchment Areas:

Number of clients receiving services : 1091

State Funded Pediatric Sickle Cell Clinics

Northern Virginia

INOVA Fairfax - Falls Church – Clients served: 204 (19%)

West

UVA - Charlottesville
Clients served: 102 (9%)

Eastern Virginia

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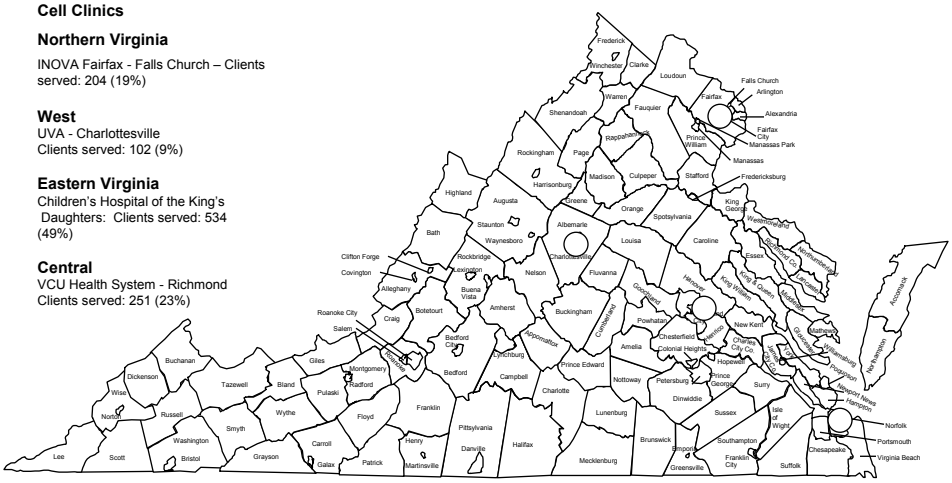


What are you paying for?

Children's Hospital of the King's Daughters Serving >500 families

- **50% Nurse Manager**
- **100% Transition Coordinator**

- **Inova Fairfax Hospital for Children Serving > 200 families**
  - **85% Nurse Manager**
- **University of Virginia Serving > 100**
  - **20% Nurse Practitioner,**
  - **25% Social Worker**
- **VCU Health System Serving >250**
  - **75% Nurse Practitioner**
  - **25% Social Worker**

Community-based Sickle Cell Programs

State and local funding

Local Sickle Cell Chapters

Local Chapter	Location
Sickle Cell Awareness Association of Central Virginia	Lynchburg
Fredericksburg Area Sickle Cell Association	Fredericksburg
Sickle Cell Association, Inc.	Norfolk
Halifax-South Boston Sickle Cell Chapter	South Boston
Blueridge Area Sickle Cell Association	Rocky Mount
Danville-Pittsylvania County Sickle Cell Association	Danville
Virginians Associated for the Relief from Sickle Cell Anemia, Inc.	Richmond
Peninsula Association for Sickle Cell Anemia, Inc.	Hampton
Community-based Support Groups	
Hope for Sickle Cell Disease	Sterling
Family and Friends Fighting Sickle Cell Anemia	Richmond
Organization for Sickle Cell Anemia Resources	Richmond

State-funded Community-based Sickle Cell Programs

2007 Budget Amendment

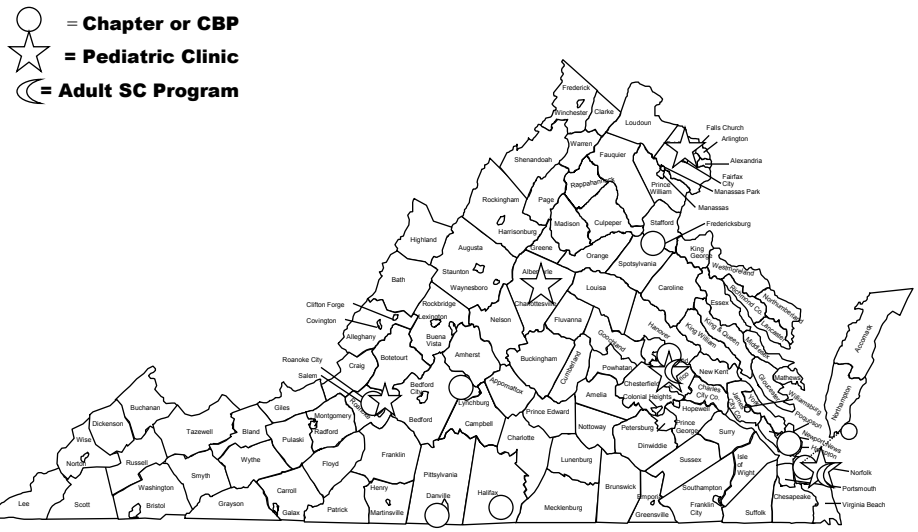
- $100,000 yearly allocation approved for grants to community-based programs that provide education and family-centered support for individuals suffering from sickle cell disease

- Allocated through a competitive RFP process
- Administered by the Department of Health
- Monitored through a yearly report to the Committee on Health and Human Services

2007 Awards reduced to $50,000 (50%)

- Sickle Cell Association of Hampton Roads
  - Norfolk
- Organization for Sickle Cell Anemia Resources
  - Richmond
- Fredericksburg Area Sickle Cell Association
  - Fredericksburg

Chapters and Funded Clinics Across the Commonwealth



Trends and Challenges

Trends

In just one generation, the average survival of patients with sickle cell anemia has increased from 14 years to nearly 50 years.

American Family Physician, 2006

- In the next 5 years, 400 adolescents will transition to adult chronic disease management
- Will the health care system be ready for them?

Vision

A comprehensive program, similar to those that exist for patients with other chronic illnesses such as Type II diabetes or cystic fibrosis, will further improve the quality of life for this very vulnerable group of patients.

Challenges

- Increased funding for the development of Adult Comprehensive Sickle Cell Clinics so that our young adults do not begin to utilize expensive emergency care
  - Currently no funding for hydroxyurea treatment for adults without insurance
  - Inequities in care among African Americans, in general, extend to this subpopulation
- Increased access to care outside of urban areas through the development of a network of “experts” who would serve as resources for community-based providers, hospitals or Federally Qualified Health Centers
- Enhance health education and awareness of sickle cell disease through schools of medicine, nursing, social work and teaching

On behalf of all who wait for a cure, thank you for your concern and continued support.

http://www.vahealth.org/sicklecell

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